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COEXISTENCE OF HYPERKERATOSIS LENTICULARIS PERSTANS (FLEGEL) AND HYPERKERATOSIS FOLLICULARIS ET PARAFOLLICULARIS IN CUTEM PENETRANS (KYRLE) IN A PATIENT

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Abstract. On the basis of the literature and of a clinical case, it is suggested that Kyrle's disease and hyperkeratosis lenticularis perstans of Flegel may be different manifestations of the same disease. While there may be classical separate forms of these two conditions, one should be on the lookout for the coexistence of the two dermatoses in the same patient, as in the case here reported.

Hyperkeratosis lenticularis perstans (HLP) (4) seems to be a clinically and histologically characteristic skin condition. Since the first report in the English literature in 1968 (6) the number of published cases has steadily increased (1, 7, 9, 11). Recently we had occasion to observe a patient with typical lesions of HLP over his feet and legs and characteristic lesions of Kyrle's disease over other regions of his skin. With this case in mind, and after having studied the histopathological changes of these two dermatoses, we wonder if they could be different manifestations of the same disease.

CASE REPORT

A man aged 64 years, a retired engineer, had had various keratotic lesions over his extremities and over his trunk for many years. In recent years he had been treated for solar keratoses. In June, 1969, he was diagnosed as having HLP over the feet and legs, and Kyrle's disease of the buttocks, back of the thighs, extensor sides of elbows and over the trunk.

There was no history of similar skin changes in other members of his family. He does not know if his only brother is still alive; his sister died at the age of 29 years, and his only son is 29 years old and does not have similar skin changes.

In 1946 the patient was diagnosed as having pulmonary fibrosis; in 1963 he had peptic ulcer and hiatus hernia, and in 1965 coronary sclerosis and cervical spondylosis.

The present skin condition started at the age of 38 years when he was in the Navy (1940–1946) as an Engineroom Artificer, which entailed work in a very hot atmosphere. The dermatosis started over the region of the ankles and later extended over the extremities and over the back. There was no itching.

At examination (1969), the skin of the extremities was dry and atrophic-looking. Over the tops of the feet and around the ankles there were numerous, irregularly-shaped, warty crusty keratoses of a somewhat psoriasiform appearance, typical of hyperkeratosis lenticularis perstans (Fig. 1). There were no changes of the nail plates. Over the palms and soles there were a few irregularly scattered pinpoint sized keratoses and some pits interrupting the papillary lines, probably results of the fallen-out keratoses (Fig. 2). Over the back of the hands and over the dorsa of the proximal phalanges of the fingers there were numerous keratoses which gave the general impression of ordinary solar keratoses, although some might have been elements of HLP. The legs and thighs, the forearms and the arms, the buttocks and the back were covered by hundreds of keratoses of varying size and shape. Many of them, especially those over the back, were very small pinhead sized, more palpable than visible. Some were filiform miniature cutaneous horns. The lesions over the extensor sides of the elbows, over the buttocks and over the back of the thighs were round elevated papules with a central round horny plug. One was able to remove this horny adherent penetrating plug in one piece, leaving behind a funnel-shaped depression, which was red and showed bleeding from its depth. The appearance of these lesions, with the greyish central round keratotic elevation surrounded by a regular round infiltrated red border, was clinically characteristic of Kyrle's disease (Fig. 3).

Over the abdomen there were two or three small papular keratoses.

The patient had Fordyce disease of the mucous membrane of the cheeks and a caviar tongue. No keratoses were visible in the mouth.

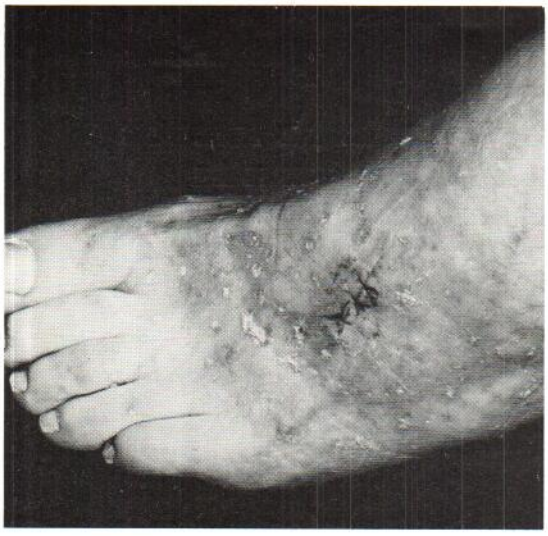

Fig. 1. Hyperkeratosis lenticularis like irregular psoriasiform keratoses of the top of the foot.

PATHOLOGY

Two biopsies of the skin were obtained; one was from the dorsum of the right foot, the other from the right buttock. The tissue was fixed in 4% formaldehyde and embedded in paraffin. Sections 5 μm thick were stained with haematoxylin and eosin, periodic acid-Schiff (PAS), alcian blue-PAS, silver impregnation method for reticulin, and by the Masson trichrome and the Verhoeff/van Gieson methods.

Skin from dorsum of foot: The histological picture is distinctive. A significant feature is the presence of a patchy massive keratosis which measures up to 850 μm in thickness and which, in places, is associated with depression and thinning of the stratum malpighii. The epidermis in these areas may be reduced to 1 or 2 cell layers. In the keratotic layer are noticed many fine linear slits which lie parallel to the surface and extend for long distances. Inconspicuous areas of focal parakeratosis are also present, particularly in the keratin layer which bulges into the epidermis. The granular cell layer is often reduced to a single cell line. The epidermis is atrophic but, in some areas, forms distinctive circumscribed cone-shaped elevations. In addition, strands of epidermis proliferate downwards into the dermis to form conspicuous irregular ridges, usually 2 cells thick and about 70 μm in length. The papillary and sub-papillary layers of the upper dermis show oedema and a mild chronic inflammatory infiltrate which is composed mainly of lymphocytes but also contains histiocytes.

The capillaries are numerous and dilated and the endothelial lining is swollen and prominent. The cellular infiltrate is most marked in the epidermal elevations and its lower margin is sharp. Other changes include fragmentation and loss of the elastic fibres in the civinity of the infiltrate and an abundance of reticulin fibres around the capillaries in the superficial layers of the dermis.

The histological picture is thus typical of hyperkeratosis lenticularis perstans. In addition it has been possible to demonstrate changes which are similar to those seen in Kyrle's disease (Figs. 4, 5 and 6).

Skin from buttock: A massive keratotic plug, which is partly parakeratotic and contains areas of basophilic degenerate material, extends deeply into the epidermis. The epidermis at the base of the invagination is compressed and, in places, only 2 or 3 layers of attenuated cells separate the horny plug from the dermis, but at no point is there full penetration of the epidermis. The

Fig. 2. Pits interrupting the papillary lines of the palms.

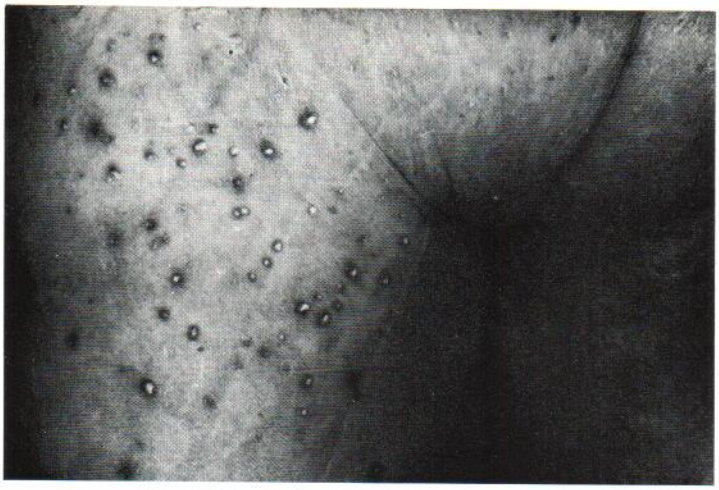

Fig. 3. Kyrle type of keratoses of the back of the thighs.

basophilic material does not give a reaction with elastic tissue stains and the parakeratosis involves almost all layers of the epidermis at some sites. The epidermis is thinned with flattening of the rete ridges, but at each margin of the massive horny thickening the epidermis shows a cone-shaped elevation with moderate acanthosis beyond this limit. Inflammatory changes are present in the uppeer dermis at the margin of the plug of keratin.

The morphological picture is typical of Kyrle's disease (Fig. 7).

COMMENT

The clinical and histopathological changes of Kyrle's disease were recently summarized by Carter & Constantine (2, 3). In our patient, the lesions over the thighs, buttocks and over the extensor sides of the elbows seem to conform with the classical changes shown by Kyrle's keratosis. The papular round elevations with their central penetrating keratotic core, which when removed leaves behind a funnel-shaped depression, are very characteristic of this dermatosis. Most of these lesions in our patient were single and scattered over the areas involved, with little tendency to confluence. In the differential diagnosis of Kyrle's disease one has to consider the recently described perforating folliculitis of Mehregan & Coskey (8). The lesions of Kyrle involve much more extensive skin areas—its keratotic papules are larger than those of perforating folliculitis. In Kyrle's disease the keratotic plugs are not necessarily of follicular location, while they are strictly

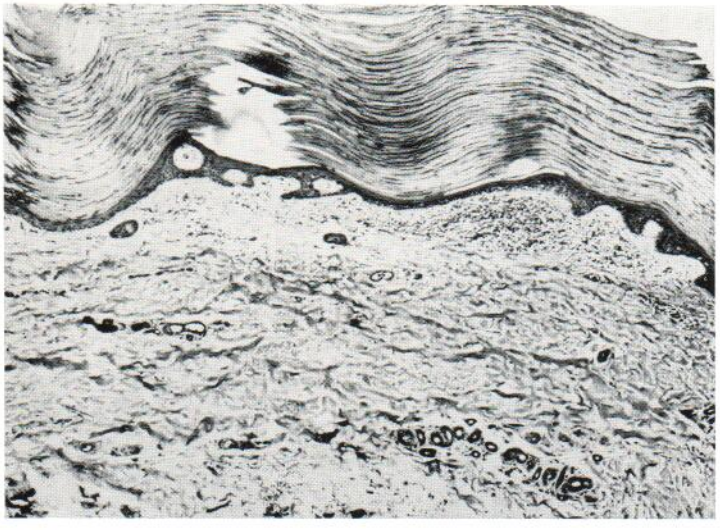

Fig. 4. Lesion from the foot. The changes include considerable hyperkeratosis with bulging into the epidermis (Kyrle-like) and a thin epidermis showing a circumscribed cone-shaped elevation on the left side. An inflammatory infiltrate with a sharp lower border in the upper portion of the dermis is seen on the right side (Flegel-like). H. and E., × 50.

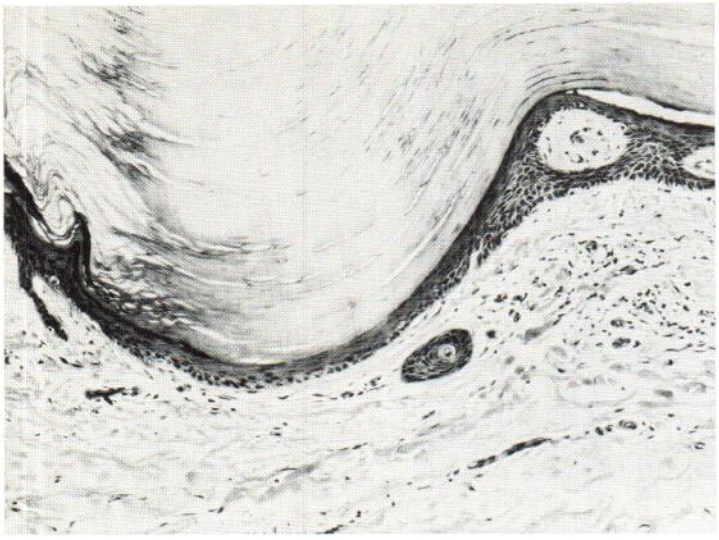

Fig. 5. High-power view of part of Fig. 1. showing depression of the massive horny layer (Kyrle-like). H. and E., × 125.

follicular in perforating folliculitis. Kyrle's disease is a rare condition, while the dermatosis of Mehregan & Coskey seems to be quite common. The psoriasiform irregular patchy superficial keratoses seen over the tops of the feet and around the ankles of our patient do not look at all like Kyrle's keratoses and have all the appearances of HLP as first reported and illustrated by Flegel (4). The dermatosis seems to have started around the ankles, as is usual for HLP.

The papular palmo-plantar keratoses and the pits interrupting the papillary lines of the palms and soles, which were first reported in HLP by Grüneberg (5) and were seen in our previous case of HLP, reported with Bear (6), were again observed in our present patient. Grüneberg considered the presence of these lesions as a possible diagnostic element in favour of acrokeratosis verruciformis or Darier's disease. As we pointed out in our previous case report, there is nothing else in the clinical or in the histopathological picture suggestive of these last-mentioned dermatoses. Interestingly enough, when summarizing the clinical characteristics of Kyrle's disease, Carter & Constantine (2) stated: "Lesions do not involve the palmar and plantar surface." We have to consider then the changes of the palms and soles as probably belonging to he picture of HLP. We do this, however, with hesitation since from a purely morphological point of view the round keratotic

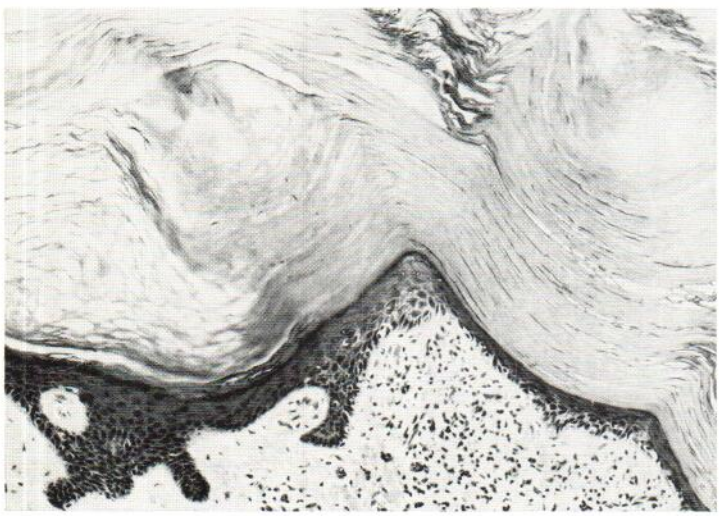

Fig. 6. Lesion of the foot showing the thick keratotic layer, a cone-shaped elevation of the atrophic epidermis, and a well-circumscribed inflammatory infiltrate in the upper dermis (Flegel-like). H. and E., × 125.

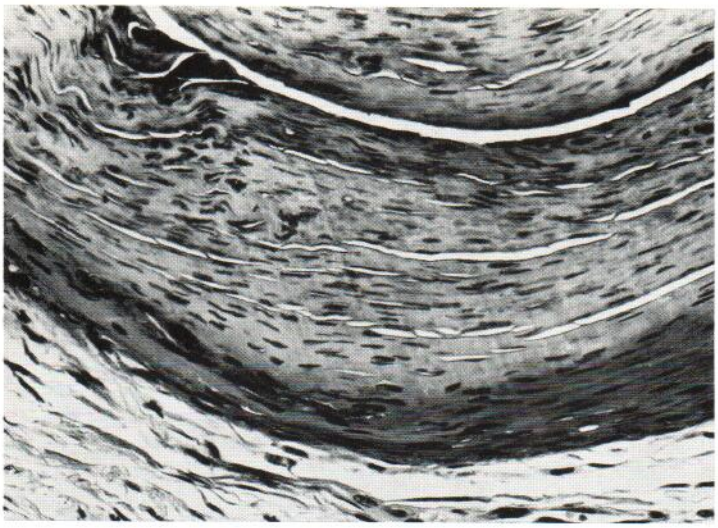

Fig. 7. Lesion from the buttock. The changes are typical of Kyrle's disease. A massive keratotic plug extends deeply into the epidermis. The parakeratotic changes are contiguous with the dermis. There is practically no basal layer. In the left upper corner is seen an area of basophilic material which is not associated with an inflammatory infiltrate; however, this degenerate material often shows an intraepidermal cellular response consisting mainly of neutrophils. H. and E., × 312.

plugs and the resulting pits are similar to the lesions of Kyrle's keratoses, and do not remind one of the irregular patchy psoriasiform keratoses of HLP.

The involvement of the trunk in HLP was first reported by us (6) and, in the case of Zina & Pippione (11), was the only region involved. The lesions of the trunk are round discrete keratotic papules, which are like the keratoses of Kyrle.

The biopsy taken from the HLP type lesion of the feet showed the characteristic changes of HLP, with areas of subepidermal infiltrate hugging an atrophic epidermis covered by a greatly thickened hyperkeratotic layer. The infiltrate, as described in the earlier reports, had a sharp lower border. There was no exocytosis into the epidermis. Associated with the histological picture which we consider diagnostic of HLP were areas of funnel-shaped depressions filled with parakeratotic laminae of horny cells, not unlike the picture of Kyrle's disease. The elastica was missing in the areas of the upper dermal infiltrate.

According to Constantine & Carter, in Kyrle's disease there is an epidermal invagination which may be of follicular or extra-follicular location and is filled by a partly parakeratotic plug. The depressed epidermis around this plug is atrophic. In some places the parakeratotic plug seems to be in direct contact with the dermis. There is a lymphocytic and histiocytic infiltrate about the lesion. The penetration of the parakeratotic plug into the upper dermis and the granulomatous reaction of the dermis to this keratotic material was considered as an essential diagnostic element of Kyrle's disease. Graciansky & Quiroga, cited by Constantine & Carter (2, 3), independently reported cases of Kyrle's disease without evidence of this penetration. In these examples, and in the fifth case of Constantine & Carter, although the parakeratotic material of the plug was in direct contact with the dermis, there was no penetration of parakeratotic material into the dermis. Our case was similar to these three previously reported ones. The presence of basophilic debris in the parakeratotic plug was considered an important diagnostic element by Constantine & Carter. This basophilic material, which was also seen in our case, was regarded by these authors to constitute in all probability the degenerated cells of the reactionary inflammatory infiltrate, with which the dermis responds to the penetrating extraneous horny material; the infiltrate is then surrounded by the epidermis and later moved upwards by the proliferating epithelium. The presence of this basophilic debris is therefore considered to be strong evidence of the actual penetration of the dermis by the horny material which has taken place previously.

In our present patient we have found clinical and histopathological evidence of the coexistence of HLP and of Kyrle's disease. Although we are able to separate the two dermatoses clinically, we have to admit that their histopathological picture shows many similarities and transitional stages. In the recent "Textbook of Dermatology" of Rook, Wilkinson & Ebling (10), two illustrations of

Kyrle's disease by E. Wilson-Jones (p. 1051) show the most characteristic changes of HLP). There is nothing in these pictures to remind one of Kyrle's keratoses and everything to make a diagnosis of HLP.

In both Kyrle's and Flegel's diseases, if they are separate diseases and not stages or regional variations of the same condition, we might be dealing with genodermatoses of late onset. This was suggested in our first paper for HLP, and since then has received support from Bean (1), who reported on the familial incidence of the condition. Kyrle's disease has also been reported in siblings.

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